Transition to Adult Care in Patients with Sickle Cell Disease

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Objectives

• Children’s Sickle Youth Network (CYSN™)
• Pathophysiology of sickle cell disease
• Common complications affecting adolescents and young adults with sickle cell disease
• Core Elements of Transition
• Findings from a qualitative study on SCD transition
• Multidisciplinary roles in transition in sickle cell disease
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Sickle cell in Alabama

• Most common hemoglobinopathy in AL
  – 1/12 African Americans with sickle cell trait
  – 1/350 African Americans are born with SCD

• Screening done by high performance liquid chromatography (HPLC) on heel stick

• Hemoglobinopathy screening started in 1988
Children’s Sickle Youth Network (CYSN™)

- Multidisciplinary team: hematologists, nurse practitioners, nurses, social workers, psychologist, school liaison

- Provides care for more than 1000 patients with sickle cell disease (SCD) in Alabama

- Manages >80 patients/month on chronic transfusion
Children’s Sickle Youth Network (CYSN™) Catchment Area
Children’s Sickle Youth Network (CYSN™) Standards of Care

- Infection control (prophylactic penicillin)
- Annual screening for risk of stroke (transcranial doppler)
- Patient/family education
- Hydroxyurea therapy
- Chronic blood transfusion therapy (UAB)
- Screening for common comorbidities: asthma, kidney disease
- Transition readiness
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What is Sickle Cell Disease?

• An autosomal recessive red blood cell disorder

• Primarily affects individuals of African descent

• Characterized by:
  o early red blood cell hemolysis - ->anemia
  o widespread inflammation
  o recurrent episodes of pain
  o risk for infection and end-organ damage
Pathophysiology of SCD

Normal Red Blood Cell  Sickled Red Blood Cell
Common SCD complications

**CHRONIC COMPLICATIONS**
- Retinopathy
- Obstructive Sleep Apnea
- Anemia, Leukocytosis
- Pulmonary Hypertension
- Cardiomegaly
- Functional Asplenia
- Indirect Hyperbilirubinemia
- Isosthenuria, Chronic Renal Failure
- Avascular Necrosis
- Delayed Puberty, Priapism
- Skin Ulcers

**ACUTE COMPLICATIONS**
- Post-Hyphema Glaucoma, Retinal Infarct
- Stroke, Meningitis
- Acute Chest Syndrome
- Sickle Hepatopathy
- Splenic Sequestration, Splenic Infarct
- Papillary Necrosis
- Cholelithiasis
- Bone Marrow Infarct, Osteomyelitis
Mortality in SCD

• 1973 autopsy review: estimated median survival of 14.3y
• Cooperative Study of Sickle cell disease (CSSCD) 1979-87 (Leikin), death rate highest in 1-3yo, probability of living past 20yo 85-95%
• Platt CSSCD follow up in 1994: median age of death 42y for male, 48y for female
• Lanzkron (2013): improved mortality in children with SCD with increasing mortality in adults 1979-2005

Diggs LM, Anatomic lessons in sickle cell disease in Sickle cell idisease: diagnosis, management, education and research 1973
Leikin SL et al Mortality in children and adolescents with sickle cell disease Pediatrics 1989
Platt et al Mortality in sickle cell disease: Life expectancy and risk factors for early death. NEJM 1994
Lanzkron Mortality rates and age at death from sickle cell disease US 1979-2005 Public health rep 2013
A Legacy of Excellence in Sickle Cell Disease Research—Extending Life Expectation

Year


Life Expectancy (Years)

0 5 10 15 20 25 30 35 40 45 50

Transfusion

Penicillin

National Sickle Cell Act

Hydroxyurea

1916
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Most common complications affecting adolescents and young adults with SCD

- Stroke
- Iron overload
- Chronic lung disease
- Pulmonary hypertension
- Chronic kidney disease
- Venous thromboembolism
Potential impact of a stroke during early adulthood

- Stroke
- Cognitive deficits
- Unemployment
- Poor access to care
- Uninsured
- Low income
- Increased morbidity
- Risk for early death
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Transition ≠ Transfer
What is Transition?

• “Purposeful, planned movement of adolescents with chronic medical conditions from child-centered to adult-oriented health care”

• Goal of maximizing “lifelong functioning and potential through the provision of high-quality, developmentally appropriate health care services that continue uninterrupted as the individual moves from adolescence to adulthood.”


Why is transition important in SCD?

• 93% of children with sickle cell disease will live 18 years old

• Increased morbidity

• Increased risk for premature mortality
Imbalance between number of adults providers and number of adults with SCD

Adults with SCD  Adult providers for SCD (Primary care, Hematologists)
Why is transition important in SCD?

- 2012 Dickerson: Young adults with sickle cell anemia have:
  - Twice the number of ER visits and inpatient stays per year than those 10-17yo or 31-45yo
  - Highest percentage of frequent acute care visits (>10 visits per year) and return to acute care within 14 days

- 2010 Quinn: Within Dallas Newborn cohort, most deaths (6/7) in patients 18-23yo occurred within 2 years of transition to adult care.


Why is transition important in SCD?

- Hamideh 2013: “Mortality rates in patients with sickle cell disease increased from 0.6 in the 15-19 year group to 1.4/100,000 in the 20-24 year group”
- Blinder 2013: “Patients transitioning to adult care received less transfusions and hydroxyurea, less iron chelation therapy, had higher healthcare costs and suffered from more frequent SCD related complications than pediatric patients.”

Hamideh. Sickle cell disease related mortality in the US 1999-2009 PBC 2013
Blinder. Age related treatment patterns in sickle cell disease patients and the associated sickle cell complications and healthcare costs. PBC 2013
SCD Quality Guidelines (Wang 2011)

Adolescents with SCD:

• Should have a “transition plan to adult care including a written transfer summary of medical history”
• Should receive counseling regarding the transition process
Health People 2020 Goal
(Dept. of Health and Human Services)

Increase the proportion of youth with special healthcare needs who’s healthcare provider has discussed transition planning from pediatric to adult care
Got Transition

• Agreement between the Maternal Child Health Bureau and the national alliance to advance adolescent health

• Advocates for six core elements of healthcare transition
Six core elements of transition

1. Transition policy
   - Develop policy
   - Educate staff on approach
   - Post policy and discuss with youth and families

2. Transition tracking and monitoring
   - Develop database and use to track progress of patients

3. Transition readiness
   - Conduct transition readiness assessments beginning at age 14
   - Develop goals and actions and document
Transition Readiness Assessment template for SCD-DISEASE KNOWLEDGE

<p>| I know what type of sickle cell disease I have. |
| I know my medical needs and can explain them to someone. |
| I know what a hematologist is and why I go to one. |
| I know what to do in case of a medical emergency. |
| I understand what causes a pain episode. |
| I understand how drugs, alcohol and tobacco affect sickle cell disease. |
| I have friends that I can talk to about sickle cell disease. |
| I know about necessary screening exams (echo annually, kidney function annually, retinal exams, etc.). |
| I know how to get blood work and x-rays. |</p>
<table>
<thead>
<tr>
<th>I know what my medications are for.</th>
</tr>
</thead>
<tbody>
<tr>
<td>I know the names and doses of my medications.</td>
</tr>
<tr>
<td>I remember to take my medications without my parent reminding me.</td>
</tr>
<tr>
<td>I fill prescriptions before I run out of medications.</td>
</tr>
<tr>
<td>I am aware of what hydroxyurea is and how it prevents sickling of my red blood cells.</td>
</tr>
<tr>
<td>I know how to prevent a pain episode and what to do if I have pain.</td>
</tr>
</tbody>
</table>
### Transition Readiness Assessment template for SCD-APPOINTMENTS

<table>
<thead>
<tr>
<th>Task</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>I make my own doctors’ appointments.</td>
<td></td>
</tr>
<tr>
<td>I know how to get medical care when the doctor’s office is closed.</td>
<td></td>
</tr>
<tr>
<td>I fill out my own medical history form</td>
<td></td>
</tr>
<tr>
<td>I keep track of my own medical information.</td>
<td></td>
</tr>
<tr>
<td>I keep track of my doctors’ and other appointments.</td>
<td></td>
</tr>
<tr>
<td>I make a list of questions before my visit with my doctors.</td>
<td></td>
</tr>
<tr>
<td>I answer questions on my own during medical visits.</td>
<td></td>
</tr>
<tr>
<td>I arrange my own transportation to medical appointments.</td>
<td></td>
</tr>
<tr>
<td>INSURANCE</td>
<td></td>
</tr>
<tr>
<td>-----------------------------------------------</td>
<td></td>
</tr>
<tr>
<td>I carry my own insurance card.</td>
<td></td>
</tr>
<tr>
<td>I understand my insurance plan.</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>PRIVACY INFORMATION</th>
</tr>
</thead>
<tbody>
<tr>
<td>I understand how health care privacy changes at age 18, when I am legally an adult.</td>
</tr>
</tbody>
</table>
Six core elements of transition

4. Transition planning
   • Develop and update plan of care
   • Prepare for adult approach to care at 18yo including legal changes
   • Determine need for decision making support
   • Plan with patient/parent timing of transfer
   • Assist youth in identifying adult provider
   • Provide insurance and self care resources
Six core elements of transition

5. Transfer of care
   - Confirm date of first adult provider contact
   - Transfer when patient’s condition is stable
   - Complete transfer package
   - Continue to see in pediatric setting until young adult is seen in the adult setting
Six core elements of transition

6. Transfer completion

- Contact young adult 3-6m post-transfer to confirm transfer and obtain feedback
- Communicate with adult practice and offer consultation assistance as needed
- Build collaborative partnerships with adult primary and specialty care providers
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EXPLORING ADULT CARE EXPERIENCES AND BARRIERS TO TRANSITION IN ADULT PATIENTS WITH SICKLE CELL DISEASE

Study Methodology

• Patients >18yo from UAB sickle cell clinic (no limit on genotype)

• Semi-structured interviews
  – Timing and preparation for transfer
  – Development of independence
  – Feelings about and experiences in pediatric and adult care

• Review and coding of transcripts by CB and JL
Patient demographics

- 10 participants
- 24-55yo
- All patients had insurance
- 9/10 on disability
- 9/10 HS graduates
## Pre-transition concerns

<table>
<thead>
<tr>
<th>Concern</th>
<th>Reference(s)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Leaving comfortable pediatric setting</td>
<td>10, 11, 14, 15</td>
</tr>
<tr>
<td>Going to a doctor unfamiliar with treating sickle cell disease</td>
<td>11</td>
</tr>
<tr>
<td>Will my parents allow patients to become independent</td>
<td>11, 15</td>
</tr>
<tr>
<td>How will I pay for medical treatment</td>
<td>10</td>
</tr>
<tr>
<td>Will I be treated as an adult</td>
<td>10</td>
</tr>
<tr>
<td>Will adult providers understand how sickle cell disease affects me as an individual</td>
<td>10, 14</td>
</tr>
</tbody>
</table>
Living with SCD as an adult

- Poor understanding of disease (non-hematologists)
- Pain management
- Co-Workers
- Family relationships/Marriage
- Physician Mistrust
- Relationships
- Access to care
- Transportation
- Finding SCD providers
- Working with SCD
- Job Termination
- Employment
- Insurance
- Loss of Insurance
- Accept patient’s insurance
- Limited Medication Refills
Living with SCD as an Adult – Themes Identified

- Physician mistrust
  - “Treat me like you would want someone to treat you. Don’t group me in a category of other SCD patients.”
  - “Dr. X is my primary doctor but he doesn’t really know anything about sickle cell disease so I have to tell him what to do.”
  - “he (Primary Medical Doctor) was really mean to me. He was a little bit arrogant. It was like he was blaming me for the sore on my leg (leg ulcers).”
  - “When you go to the emergency room...they think its an opinion but its not, it just hurts.”
  - “They...treated us like people off the street. Oh, she’s just coming in here to get a fix. The worst thing is that they don’t treat you with respect and kindness. They treat you like a crackhead because of what you have.”
  - “When you get to be an adult, they tend not to believe you as much.”
Living with SCD as an Adult – Themes Identified

- Employment
  - “You can’t go in there telling them that you have these crises and I have to be out”.
  - “It’s very difficult to hold employment because of the stress level.”

- Relationships at work and at home:
  - “where have you been, on vacation?”
  - “(he did not) understand the cycle of pain and what it does.”
  - “If I say ‘oh my god my leg hurts so bad’ than he’d say ‘well mine hurts too’ I just say OK dad, but it’s a different type of hurt.”
Patient and family fears:

• Leaving the current provider
  o “It was like someone pulled the rug out under me”

• Establishing trust with a new provider
  o “Who was going to be my doctor and was she going to be nice from the time when I come over?”

• Navigating a new health care system
  o “I was definitely concerned about switching because that’s all I knew—Children’s and the doctors I was seeing there. I was definitely scared to move over to UAB”
Conclusions

• Patients perceive significant distrust from physicians
  • Interventions to improve physician opinions regarding pts with SCD
• Continued parent involvement
  • May be used as a transition
• Lack of physician knowledge
  • Improved education efforts
Conclusions

• Difficulty identifying doctors
  o Appropriate care paradigm?
• Difficulty maintaining employment
  o Further research needed
• Questions regarding numbers of patients with sickle cell
  o 2 pts diagnosed as adults
  o 2 pts with no healthcare contact for many years
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Factors that influence success of transition to adult life

- Family/Relationships
- Health
- Transition
- Education
- Vocation
- Life skills
- Self-efficacy
Transition Process

- **<12yo**
  - Encourage parents to teach children about their disease including good health practices

- **12-14yo**
  - Begin encouraging patients to learn more about their disease

- **15-18yo**
  - Patient transition planning and education

- **>18yo**
  - Transfer to adult care
Transition Process

• Engage patients in the conversation regardless of age
• Make every clinic visit an educational visit
• Follow school progress to ensure staying at appropriate grade level
• Encourage dreams while maintaining a focus on goals within reach
Important things to know

<19yo

- Medicaid eligibility based on parental income
- Unlimited inpatient and outpatient visits
- Dental services
- Yearly eye exams and eyeglasses every 2 years
- Parking is free or validated for patients/parents at COA for appointments
- COA social workers/ KID 1 will provide transportation assistance.

Adulthood

- Unless disabled, no longer eligible for Medicaid
- Limited to 14 combined outpatient and inpatient visits
- No dental services
- Eye exams every 3 years, no eyeglasses
- No free parking
- No KID 1 or transportation assistance
Transition resources

• www.gottransition.org
• http://www.sickkids.ca/Good2Go/index.html
• http://hctransitions.ichp.ufl.edu/hct-promo/
Questions?